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A pulmonary arteriovenous shunt hypoxemia and recurrent bleeding in the emergency department: a case report

Sonia Zotti,[1] Dino Eminente,[1] Silvia Navarin,[1] Tommaso Grandi,[1] Federica Sambuco,[1] Gian Paolo Ussia,[2] Francesco Travaglino[1]

[1]Emergency Department, Fondazione Policlinico Universitario Campus Bio-Medico di Roma; [2]Hemodynamics and Interventional Cardiology Unit, Fondazione Policlinico Universitario Campus Bio-Medico di Roma, Italy

Correspondence: Dino Eminente, Emergency Department, Fondazione Policlinico Universitario Campus Bio-Medico di Roma, Italy. E-mail: dino.eminente@unicampus.it

Key words: Rendu-Osler-Weber syndrome; hereditary hemorrhagic telangiectasia; pulmonary arteriovenous malformations.

Abstract

We report the case of a 62-year-old male with a long-standing history of Hereditary Hemorrhagic Telangiectasia (HHT, Rendu-Osler-Weber syndrome) complicated by multiple Pulmonary Arteriovenous Malformations (PAVMs), chronic anemia from recurrent gastrointestinal bleeding, and permanent atrial fibrillation. He presented to the Emergency Department (ED) with acute dyspnea and atrial fibrillation with rapid ventricular response. Despite high-flow oxygen therapy, severe hypoxemia persisted due to right-to-left shunt physiology. A prompt blood transfusion increased the oxygen delivery, contributing to transient clinical stabilization; however, the PaO_2/FiO_2 ratio remained severely reduced, confirming persistent impairment of oxygenation. After multidisciplinary evaluation, he underwent a successful embolization of one PAVMs, with overall clinical improvement. Because of high bleeding risk, long-term oral anticoagulation was avoided and left atrial appendage closure was proposed but declined by the patient. This case highlights the complexity of managing shunt hypoxia and anticoagulation in HHT patients with PAVMs presenting with acute dyspnea to the ED.

Introduction

In the fast-paced environment of emergency medicine, few clinical challenges are as urgent or as complex as Acute Respiratory Failure (ARF). Whether caused by pulmonary, cardiac, or neuromuscular compromise, ARF can rapidly progress to life-threatening decompensation if not managed swiftly and effectively. Hereditary Hemorrhagic Telangiectasia (HHT) is an autosomal dominant vascular disorder characterized by mucocutaneous telangiectasias and Arteriovenous Malformations (AVMs) in multiple organs, including the lungs, the Gastrointestinal (GI) system, the liver, and brain.[1-2] Pulmonary AVMs, present in up to 50% of patients, may cause significant right-to-left shunting and severe hypoxemia refractory to supplemental oxygen.[3-4] Management of hypoxia due to HHT patients is particularly challenging when concomitant atrial fibrillation necessitates anticoagulation, given the elevated bleeding risk associated with mucosal and visceral telangiectasias.[5]

Case Presentation

A 62-year-old man with known HHT since 2006 presented to the emergency department with left periorbital edema, acute dyspnea and tachycardia. His medical history included dilated cardiomyopathy with mild-to-moderate mitral and tricuspid valve insufficiency, a previous ischemic stroke with hemorrhagic transformation in the left rolandic region (1992), chronic anemia from recurrent GI bleeding, prior pulmonary AVM embolization (2007–2009), and chronic hypoxemic respiratory failure requiring oxygen therapy. He was known to have permanent atrial fibrillation on rate control without chronic anticoagulation due to previous major bleeding events.

On admission, he was tachycardic (140 bpm) and hypoxemic (SpO_2 84% on high-flow oxygen, 60%). Arterial blood pressure was 105/60 mmHg and arterial blood gas analysis without oxygen supply showed pH 7.46, $paCO_2$ 33 mmHg, paO_2 47 mmHg, and hemoglobin 8.3 g/dL. Anaphylaxis was rapidly ruled out since the periorbital edema was linked to orbital telangiectasias known from the history taking. The severe respiratory failure and atrial fibrillation rhythm with a high ventricular rate were likely to be a case of acute decompensated heart failure. Chest X-ray reported signs of bilateral hilar congestion and basal pleural effusion, larger on the left side, and multiple scattered nodular opacities, consistent with known pulmonary AVMs and metallic coils corresponding to prior embolization of other malformations. Bedside echocardiography showed a left ventricle at the upper limits of normal size with mild concentric left ventricular hypertrophy and a global systolic function at the lower limits of normal, without clear regional wall motion

abnormalities (LVEF 51%). Mild-to-moderate functional mitral and tricuspid regurgitation with early signs of pulmonary hypertension (estimated sPAP 45 mmHg, TR Vmax 2.65 m/s) were also detected as well as dilated right heart chambers (RV basal diameter 53 mm), with right ventricular systolic function at the lower limits of normal (TAPSE 18 mm, tricuspid S' 12 cm/s). Indeed, a dilated inferior vena cava with poor inspiratory collapse was observed. In the emergency department, despite optimized High-Flow Nasal Cannula (HFNC), use of diuretics and rate-control therapy for atrial fibrillation, hypoxemia and high ventricular rate persisted. Arterial blood gas analysis on HFNC with 60% of fraction of inspired oxygen (FiO_2) was comparable to the previous analysis as pH 7.43, $paCO_2$ 39 mmHg, paO_2 58 mmHg, and hemoglobin 8.9 g/dL. As shown in Figure 1, an angio-CT-scan was then performed confirming the diffuse pattern of multiple bilateral pulmonary vascular ectasias, several of which had been previously embolized and some showing a slight increase in size compared with the prior CT, particularly those in the apical segment of the right upper lobe (18 mm *vs*. 14 mm) and in the medial segment of the middle lobe (32 mm *vs*. 27 mm). Due to the severe hypoxia without the need of intensive care unit for invasive mechanical ventilation or advanced organ support and according to our institutional guidelines, the patient was admitted to the high dependency unit of the Emergency Medicine department to undergo a specialist evaluation regarding Rendu-Osler-Weber syndrome and treat his high-rate atrial fibrillation. Anticoagulation was not started due to recurrent bleeding from his orbital telangiectasia. However, multiple transfusions of packed red blood cells were necessarily carried out to raise hemoglobin's level that was chronically low. Transfusion slightly improved the clinical condition through an increased arterial oxygen content, despite the persistent shunt that limited oxygenation, as arterial blood gas analysis on HFNC (FiO_2 80%) was pH 7.51, $paCO_2$ 31 mmHg, paO_2 70 mmHg, Lac 13 mg/dL, sO2 97.1%, HCO3- 24.7 mmol/L.

During hospitalization, after multidisciplinary evaluation, he underwent right and left heart catheterization (mean PAP 21 mmHg, wedge 20 mmHg, high cardiac output and cardiac index 13.30 L/min and 7.11 L/min/m^2) and a successful percutaneous embolization of the large AVM at the level of the right middle lobe. It had a dual afferent arterial vessel (pre-bifurcation diameter 10 mm) draining into a large, tortuous, ectatic vessel, and subsequently into the right inferior pulmonary vein. The 18–14 mm Amplatzer Vascular Plug device was positioned in the afferent vessel at the bifurcation site in a such a way that the feeding arteries were completely excluded and follow-up angiography confirmed complete exclusion of the large AVM. Post-procedure, his oxygenation improved (PaO_2 67 mmHg on venturi mask with 50% FiO_2), heart rate was successfully controlled, and he was progressively weaned to low-flow oxygen (FiO2 32% at discharge). Finally, left atrial appendage closure was offered to the patient to mitigate

thromboembolic risk without the need for chronic anticoagulation, but the patient elected not to undergo the procedure. Despite a CHA_2DS_2-VASc Score for atrial fibrillation stroke risk of 3, given the patient's history of major bleeding and severe chronic anemia, the long-term anticoagulation strategy required careful evaluation.[6] After cardiology consultation, an elective gastrointestinal endoscopy was scheduled to investigate potential bleeding sources before initiating a Direct Oral Anticoagulant (DOAC). Pending completion of the gastrointestinal work-up and definitive bleeding risk reassessment, the patient was discharged on therapeutic low-molecular-weight heparin as a temporary anticoagulation strategy, with close cardiology and hematology follow-up.

Discussion

This case exemplifies the severe hypoxemia caused by right-to-left shunt in HHT-associated pulmonary AVMs, which may not respond to oxygen therapy alone. In this condition, increasing the fraction of inspired oxygen has limited effect, since part of the blood bypasses the alveoli and is not oxygenated at all. In such patients, maintaining adequate hemoglobin levels is critical to optimize oxygen delivery. Blood transfusions can temporarily improve the clinical situation by increasing arterial oxygen content despite persistent shunting. The coexistence of atrial fibrillation adds further complexity. While anticoagulation is essential to prevent thromboembolic events, it may exacerbate life-threatening bleeding in HHT. In this case, left atrial appendage closure would have provided an alternative to chronic anticoagulation, balancing stroke prevention with bleeding control.

Conclusions

Hereditary hemorrhagic telangiectasia with pulmonary AVMs can cause profound shunt-dependent hypoxemia refractory to oxygen therapy. In this case, blood transfusion may improve oxygen delivery but it is not sufficient to ameliorate oxygenation. In patients with atrial fibrillation and high bleeding risk, left atrial appendage closure represents a viable alternative to long-term anticoagulation. Multidisciplinary care remains essential for optimizing outcomes.

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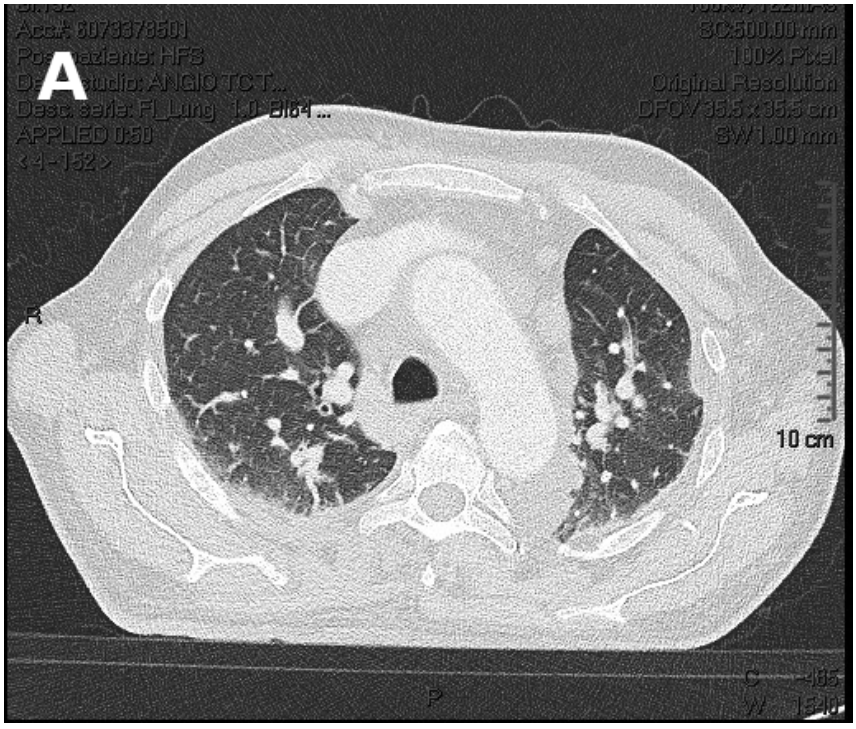
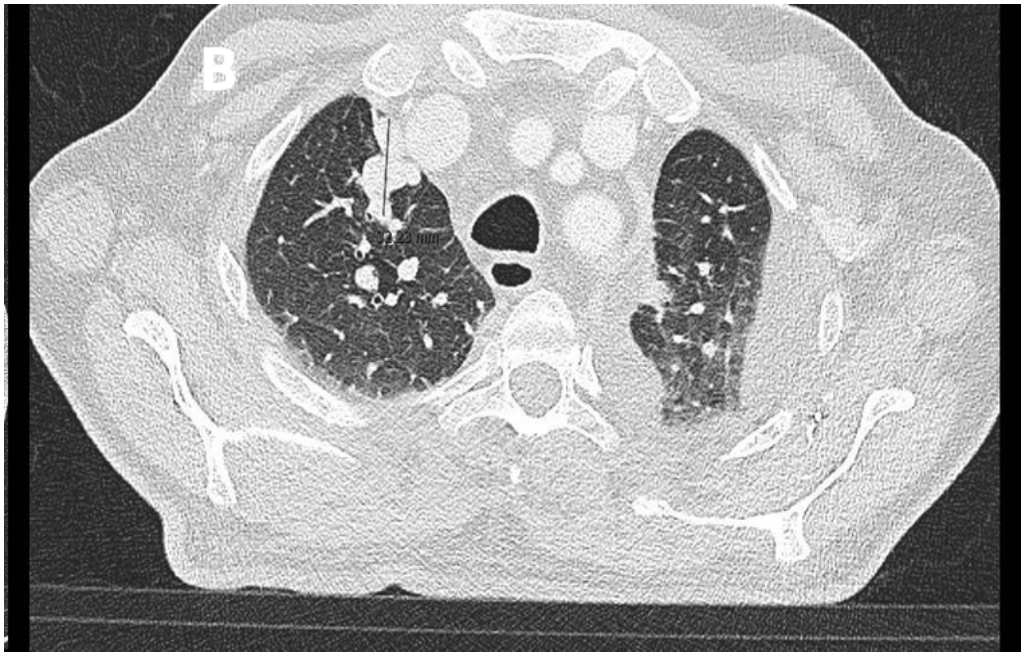
Figure 1. Chest CT angiography showing a diffuse pattern of multiple pulmonary vascular ectasias bilaterally distributed (A) and the large AVM at the level of the right middle lobe with a maximum diameter of 32 mm (B).





Contributions: the authors contributed equally to the present paper.

Ethics approval and consent to participate: no ethical committee approval was required for this case report by the Department, because this article does not contain any studies with human participants or animals. Informed consent was obtained from the patient included in this study.

Patient consent for publication: the patient gave his written consent to use his personal data for the publication of this case report and any accompanying images.

Availability of data and materials: all data underlying the findings are fully available.